Eur J Vasc Endovasc Surg **20**, 201–203 (2000)
doi:10.1053/ejvs.2000.1133, available online at http://www.idealibrary.com on IDEAL®

CASE REPORT

Severe Hypertension and Childhood Epilepsy in a 23-year-old Caucasian Woman

H. A. Khwaja[1], A. Giudiceandrea[1], P. Sweny[2] and G. Hamilton*[1]

[1]*Academic Department of Surgery and* [2]*Department of Nephrology, Royal Free Hospital, Pond Street, London NW3 2QG*

Introduction

A 23-year-old Caucasian woman was referred to the Vascular Unit at the Royal Free Hospital, London in March 1998 for further evaluation of her hypertension, initially noted during her first pregnancy in 1993. This was controlled with labetolol; however, postpartum she remained hypertensive after discontinuing the drug (BP = 170/118 mmHg). In addition, on abdominal examination a left renal bruit was found. A renal angiogram in 1996 demonstrated a 55% stenosis of the left proximal renal artery. No pressure readings were available. Renal artery angioplasty was performed on two occasions in 1996, with no effect on her hypertension. Past medical history included frequent childhood epileptic fits and curiously both her children had previous strokes, one at 9 months and the other at 14 months of age, and have since suffered from epilepsy. On admission to the Royal Free her BP was well controlled on methyldopa at 150/70 mmHg. General and cardiorespiratory examinations were normal. Abdominal examination revealed a left renal bruit but was otherwise normal. Neurological examination was unremarkable.

Baseline renal function on admission was normal (urea 2.7 mmol/l, creatinine 74 mmol/l). Inflammatory markers were also normal as was an autoimmune screen with the exception of positive antimicrosomal antibodies (titre: 1/100). Repeat renal angiography demonstrated a 50% stenosis of the proximal left renal artery and a normal right renal artery. There was also a 50% stenosis of the coeliac axis origin and a 25–50% stenosis of the superior mesenteric artery origin. Her abdominal aorta was reduced to 12 mm in transverse diameter below the mesenteric vessels (Fig. 1). A MAG-3 scan demonstrated that her right kidney contributed 58% and her left kidney 42% of total renal function and a kidney GFR estimation was 87 ml/min (normal 100–160 ml/min). Renal sizes, as determined from her abdominal CT, showed the right kidney to measure 10 cm and her left kidney 9 cm. Renal vein renin sampling indicated lateralisation to the left renal vein (1.70 pmol/ml/hr) (right renal vein 1.00 pmol/ml/hr,

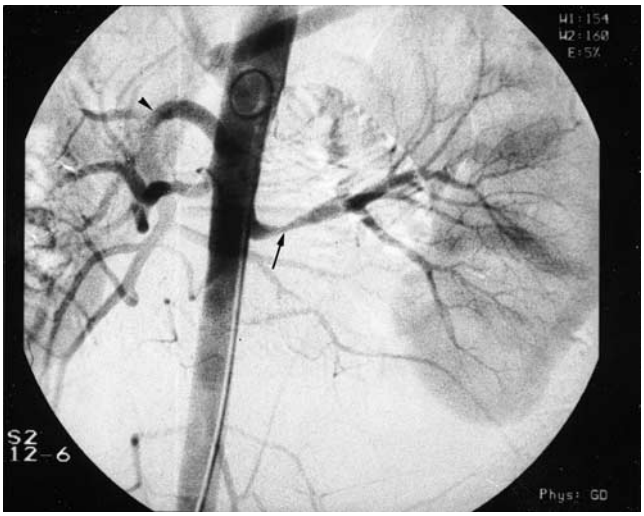

Fig. 1. Renal angiogram demonstrating stenoses of the coeliac axis, superior mesenteric artery (arrowhead) and left renal artery (arrow) as well as the abdominal aorta.

* Please address all correspondence to: G. Hamilton, Consultant General/Vascular Surgeon, Academic Department of Surgery, Royal Free Hospital, Pond Street, Hampstead, London NW3 2QG, U.K.

1078–5884/00/080201 + 03 $35.00/0

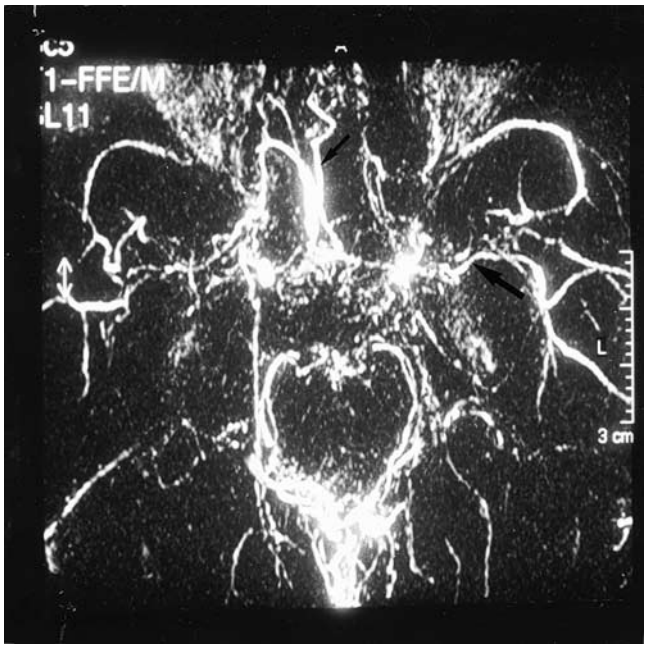

Fig. 2. Magnetic resonance cerebral angiogram demonstrating stenoses of the anterior cerebral artery (thin black arrow) and middle cerebral artery (thick black arrow) resulting in a well-developed collateral vascular system.

upper IVC 1.80 pmol/ml/h, lower IVC, 1.00 pmol/ml/h).

Carotid duplex studies demonstrated diminished sizes of both common, external and internal carotid arteries with no evidence of flow disturbances. Magnetic resonance cerebral angiography showed severe bilateral stenosis of the supraclinoid internal carotid arteries with extension into the anterior and middle cerebral arteries (Fig. 2). An extensive system of collaterals was reperfusing the peripheral branches of the anterior and middle cerebral arteries. MRI brain demonstrated areas of infarction in right middle cerebral artery territory and smaller peripherally located ischaemic lesions in the subcortical white matter of both cerebral hemispheres.

Discussion

A diagnosis of moyamoya disease was made in this patient, on the basis of the reduced diameter of her carotid and cerebral arteries. In addition, the presence of an extensive collateral vessel network from the Circle of Willis resulting in the typical "puff of smoke" appearance of these vessels on cerebral angiography confirmed the diagnosis. This patient was unique in that she had multiarterial involvement, and her lower abdominal aorta, coeliac, superior mesenteric and left renal artery were all stenosed. Interestingly, the explanation of why her children have suffered strokes at such young ages may be due to moyamoya disease. Indeed, cerebral angiography of the older child has confirmed the presence of the characteristic moyamoya circulation (personal communication, Dr V. Ramesh). In addition, the epileptic fits our patient suffered from as a child are likely to be due to areas of cerebral microinfarction secondary to moyamoya. This indeed was the explanation for why her children suffered from epilepsy.

Moyamoya disease is a rare cerebrovascular disorder of unknown aetiology, initially reported in Japanese literature in 1956.[1] The disease commonly afflicts children under 10, with a smaller peak in the 30–40 year age group.[2] The incidence in females is greater than in males (1.7:1) and there is a familial occurrence in 7% of cases.[2,3] Though primarily a condition affecting Oriental populations, there are rare reports of the disease in Caucasian patients.[4] Moyamoya is Japanese for "puff of smoke", indicative of the hazy or vague appearance of the Circle of Willis and the collateral vascular system on cerebral angiography. It is associated with fibromuscular dysplasia of systemic vessels as shown by histopathological examination of these vessels.[5] Associations of this disease with renal artery stenosis are well described.[5–7]

Surgical revascularisation procedures, i.e. extracranial–intracranial anastamoses, though effective in paediatric moyamoya disease, are of unproven value in adult cases.[8] Nevertheless, if the disease presents in adulthood the prognosis is better, though 50% of patients develop slow cognitive impairment and some may suffer from subarachnoid or intracerebral haemorrhages.[9]

It is clear from the poor results of renal angioplasty in this young lady that she has a very fibrotic left renal artery. Clinical experience of congenital renal artery stenotic disease where fibrosis is present (such as in the middle aortic syndrome) demonstrates that angioplasty not only has poor results but also has an increased risk of renal artery rupture. Renal artery stenting would not diminish these risks, and is unlikely in the presence of a fibrotic lesion to give a significantly better clinical result. Indeed, there is also the risk of restenosis. Medical management of this patient's hypertension in this situation carries a lesser risk. The patient continues to be monitored carefully. In the presence of worsening hypertension refractory to medical treatment or renal failure, the option of surgical revascularisation in this young patient will be considered.

References

1 Kudo T, Takayama R, Mikawauchi K *et al.* Occlusion of the internal carotid artery. *Proceedings of the Fifteenth Annual Meeting of Japan Neurosurgical Society* (In Japanese) 1956.
2 Fukui M. Current state of study on moyamoya disease in Japan. *Surg Neurol* 1997; **47**: 138–143.
3 Kitihare T, Ariya N, Yamaura A, Makine H, Maki Y. Familial occurrence of moyamoya disease: report of three Japanese families. *J Neurol Neurosurg Psychiatry* 1979; **42**: 208–214.
4 Yonekawa Y, Ogata N, Kaku Y, Taub E, Imhof HG. Moyamoya disease in Europe, past and present status. *Clin Neurol Neurosurg* 1997; **99** Suppl 2: S58–60.
5 Nakano T, Azuma E, Ido M *et al.* Moyamoya disease associated with bilateral renal artery stenosis. *Acta Paediatr Jpn* 1993; **35**: 354–357.
6 Van der Vliet JA, Zeilstra DJ, Van Roye SF, Merx JL, Assmann KJ. Renal artery stenosis in moyamoya syndrome. *J Cardiovasc Surg (Torino)* 1994; **35**: 441–443.
7 Choi Y, Kang BC, Kim KJ *et al.* Renovascular hypertension in children with moyamoya disease. *J Paediatr* 1997; **131**: 258–263.
8 Ueki K, Meyer FB, Mellinger JF. Moyamoya disease: The disorder and surgical treatment. *Mayo Clin Proc* 1994; **69**: 749–757.
9 Houlston RS, Saadek IK, Barker S, Hughes PJ, Lawton NF. Cerebral infarction due to moyamoya disease in an 18 year old female. *Postgrad Med J* 1993; **69**: 932–933.

Accepted 29 February 2000